

Strategies for the Management of Exstrophy, Nonpalpable Testis, Hydronephrosis

*Highlights From the Annual Meeting of the American Academy of Pediatrics Section on Urology
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The annual meeting of the American Academy of Pediatrics Section on Urology consisted of 16 sessions covering almost all areas of pediatric urology, including reconstruction, neurogenic bladder, reflux and infection, obstructive uropathy, exstrophy, hypospadias, testis, and oncology. There were 140 abstracts presented. During the meeting, which was well attended by pediatric urologists from the United States, Canada, and abroad, Bernard Churchill, MD, presented the Pediatric Urology Medal to Robert D. Jeffs, MD. Dr Jeffs received accolades for his many innovative contributions to the surgical reconstruction of bladder exstrophy. Highlights of the meeting are presented below.

Urinary Diversion

Kafer and associates¹ reviewed the Indiana University experience with the development of stomal stenosis following continent urinary diversion. In 92 patients, they utilized the appendix (n = 46), bladder wall (n = 22), or the Monti technique using reconfigured ileum (n = 14) and tapered ileum (n = 10). Of the 19 patients who developed

stomal stenosis, 58% did so within the first postoperative year. The appendicovesicostomy or bladder wall technique (constructed as a continent vesicostomy) was associated with a greater incidence of stomal stenosis than the Monti ileovesicostomy. In this series, there were no stenoses when the Monti procedure was employed.

Neurogenic Bladder

Closure of the neural tube in utero with the goal of improving neurologic function has recently been employed in human fetuses with a prenatal diagnosis of myelomeningocele.² At Vanderbilt University, 25 fetuses have undergone this innovative procedure that decreases the exposure of the spinal cord to amniotic fluid. Sixteen patients (mean age, 6.5 months) underwent urologic evaluation and were compared with myelodysplastic infants who had not undergone fetal intervention. Sonography revealed upper tract dilation in 2 patients, and voiding cystourethrography showed reflux in 2 patients. Almost half of the patients (43%) had an areflexic bladder, while 6% and 19% showed uninhibited detrusor contractions and decreased compliance, respectively. Most of the patients had a normal bladder capacity for age, but 31% had a diminished bladder capacity. Two pa-

tients required clean intermittent catheterization (CIC), and 1 was placed on anticholinergic medication. One patient experienced a significant urinary tract infection. These preliminary observations do not suggest improved lower urinary tract function following closure of the neural tube.

Exstrophy

There have been many new studies in patients with bladder exstrophy. One report from the group at Johns Hopkins University focused on patients with bladders that were too small for primary closure at birth.³ Of 605 patients with bladder exstrophy, 20 had delayed closure after 6 months (mean, 1.2 years), because the bladder template was small. Most of the patients were male (18), and osteotomies were performed in 17.

Forty-five percent achieved continence after bladder neck reconstruction, and 25% required augmentation cystoplasty and CIC. The authors concluded that a small bladder template has the potential to grow sufficiently during the first year of life. When this occurs, bladder neck plasty, augmentation cystoplasty, and ureteral reimplantation can be readily performed using the template.

Several groups recently published their experience with the Mitchell

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technique of a one-stage repair of bladder exstrophy that involves total disassembly of the epispadiac penis.⁴ Complete penile disassembly facilitates primary repair of bladder exstrophy, because it allows the bladder neck and urethra to be positioned posteriorly within the pelvis. Therefore, the proximal urethra is placed within the pelvic diaphragm in a position that is more normal anatomically.

Coplen⁵ reported on his experience with this technique in 6 newborns. The 5 neonates who had bladder closure within 24 hours did not undergo pelvic osteotomy. All of the patients had successful closure. Penoscrotal hypospadias developed in 1 patient; this was caused by urethral shortening following primary closure. One epispadiac fistula required closure. Following suprapubic tube removal at 4 weeks, all patients voided. Bladder capacities within 2 years in the 5 oldest children were 60 to 150 mL. Although there was no hydronephrosis, urinary tract infections with fever developed in 3 patients, and 1 patient required ureteral reimplantation for recurrent infections.

Kropp and Cheng's⁶ technique of total urogenital complex mobilization for the management of bladder exstrophy in female patients is based on the premise that the urethra and vagina should be treated as a single unit. To do this, complete disassembly of the pelvic floor musculature and repositioning of the urethral plate and vagina are performed. This permits the vagina to move from an anterior displaced position to a more normal perpendicular position in the perineum. To accomplish this, the pelvic floor lateral to the urethral plate and vagina must be completely dissected. A deep incision is made through the pelvic floor to the level of the anterior rectal wall. The pelvic floor musculature and pubis are then reapproximated in the midline, anterior to the urogenital complex.

Over the past 2 years, 6 female pa-

tients have undergone reconstruction using this technique. This group included 3 patients with bladder exstrophy (1 newborn and 2 older children), 2 older children with cloacal exstrophy, and 1 older child with primary epispadias. This procedure resulted in an anatomically correct position of the urogenital complex. Stenosis did not develop in the vagina of any patient. Ongoing follow-up will be needed to assess the technique's long-term effect on urinary continence.

The Johns Hopkins group⁷ reported on factors that affect the development of continence in patients with bladder exstrophy. Fifty-seven patients had undergone bladder neck reconstruction (BNR) and had adequate follow-up of more than 1 year. BNR was performed at an average age of 4.4 years, with a mean bladder capacity of approximately 90 mL. Almost 75% of the patients were continent and voiding spontaneously without intermittent catheterization. An additional 16% were dry for more than 3 hours during the day. Six patients were incontinent after BNR, and 3 have undergone continent diversion. Two important factors in achieving continence after BNR were (1) the development of a bladder capacity of 75 mL or greater, and (2) a 21-mL per year increase in bladder capacity. Only 1 patient was incontinent using these criteria.

The Johns Hopkins group⁸ also reviewed their 10-year experience with the modified Cantwell-Ransley epispadias repair. This technique was per-

formed on 93 male patients (79 with exstrophy, 14 with epispadias alone). Sixteen of the patients were undergoing secondary repair. Ten of the patients had epispadias repair at the time of bladder reclosure. Of these patients, 87 of 93 had a horizontal or downward angled penis at a mean of 68 months. Most (88 of 93) voided per urethra. There was a 23% incidence of fistulas, and 19% required a definitive closure. Strictures at the proximal anastomosis developed in only 7 patients. The urethra could be easily catheterized or easily negotiated at the time of cystoscopy in about 80% of patients.

Reflux

It is postulated that bladder spasms may be related to increased prostaglandin synthesis via cyclooxygenase (COX) pathways by sensitizing capsaicin-sensitive C-fiber neural afferents. Park and colleagues⁹ studied the effect of ketorolac, a COX inhibitor, on postoperative bladder spasms. They performed a double-blind, randomized study on patients who underwent intravesical reflux surgery. The mean age of both study groups was approximately 6 years. All patients were maintained on an epidural infusion of bupivacaine (0.1%) with fentanyl (2 µg/mL). Study patients received either ketorolac (0.5 mg/kg) or placebo at the end of the surgery and every 6 hours thereafter. The patients' families recorded the number of spasms and the degree of severity. Fewer patients experienced bladder spasms in the treat-

Main Points

- Monti ileovesicostomy can result in fewer incidents of stomal stenoses, compared with appendicovesicostomy or continent vesicostomy constructed from the bladder wall.
- Disassembly of the penis facilitates primary repair of bladder exstrophy.
- Follow-up has increased importance when a nonoperative approach to prenatally diagnosed hydronephrosis is chosen.
- Bladder spasms may be improved with ketorolac, a cyclooxygenase inhibitor.

ment group than in the control group (25% vs 83%). The mean number of bladder spasms was 1.5 in the treatment group versus 5.9 in the control group, and spasms were less severe. There were no adverse effects from the ketorolac.

Undescended Testis

Casale and coworkers¹⁰ reexamined the cost-effectiveness of laparoscopy versus initial inguinal exploration for a nonpalpable testis (NPT), because only 27% of NPT are located intra-abdominally. A prospective, randomized study was performed with 12 boys undergoing laparoscopy and 12 undergoing initial inguinal exploration for NPT. Inguinal exploration revealed atrophic testes in 58%, and laparoscopy did so in 67%. When inguinal exploration was performed first, only 1 patient (8.3%) required laparoscopy. When laparoscopy was performed first, almost all patients required inguinal exploration. The surgeons reported a mean savings of 27 minutes in the operating room and a mean cost reduction of \$2754 in operating room expenses per patient.

Lee and associates¹¹ examined whether the pretreatment location of a unilateral undescended testis influenced future fertility. They found that there was no difference in paternity rates based on pretreatment testicular location.

Hurwitz¹² studied the size of the contralateral testis in boys with unilateral NPT. It has been shown by Koff that a testis more than 2 cc in volume and more than 2 cm in length predicts monorchia in boys with NPT. Hurwitz found that 16 patients had compensatory hypertrophy of the testis (more than 2 cm) and 14 (87%) of 16 had monorchia. Two patients (12.5%) had an intra-abdominal testis, and 1 patient had a small ovotestis. A high percentage of blind-ending vasa and vessels were found, and laparoscopy was felt to be helpful. Therefore, if the testis was greater than 2 cm in length,

one could predict monorchia in 87.5%. When the testis was greater than 1.9, 1.8, or 1.7 cm in length, the predictive values were 92%, 89%, and 90%, respectively.

Stones

Noe¹³ studied the stone recurrence rate in 29 children with hypercalciuria and a history of calcium oxalate stones. No metabolic abnormality existed in any of the children. The recurrence rate for stone formation was 31% (9 of 29). Stones recurred after 1 to 15 years (average, 7.2 years). No patient at the time of stone recurrence was following the dietary and fluid recommendations suggested to them previously.

Hydronephrosis

Ulman and colleagues¹⁴ have been longtime proponents of nonoperative management of prenatally diagnosed hydronephrosis. They reported on 104 newborns with unilateral grade 3-4 hydronephrosis. All were followed nonoperatively until renal deterioration occurred, which necessitated pyeloplasty. Hydronephrosis resolved (69%) or improved (31%) in 81 infants after a mean of 78 months of follow-up. The remaining 23 infants showed an increase in hydronephrosis or a decrease in the differential on renal scan before 18 months of age. This study shows the importance of follow-up when the nonoperative approach is taken.

An alternative to surgery has been proposed following a report of the development of neurogenic bladder after bilateral megaureter reimplantation. Because of this, Kohler and colleagues¹⁵ examined the outcome of 13 patients with primary obstructive megaureter (9 bilateral, 4 unilateral) whose megaureter was managed with a double-J ureteral stent. The diagnosis was made prenatally in 12 of 13, and in 1 following a urinary tract infection. The stents were placed through an open procedure if there was an increase in hydronephrosis, a

urinary tract infection, or a decrease in renal function. The stents were placed for approximately 11 months. After at least 3 years of follow-up, 7 patients have needed no surgery, while 5 have undergone ureteral reimplantation. This study suggests that stents placed for a primary obstructed megaureter may obviate the need for ureteral surgery in more than 50% of children. ■

References

1. Kaefer M, Rink RC, Cain MP, Casale AJ. Stomal stenosis: is ileum the ideal substrate for efferent limb construction? [abstract]. *Pediatrics*. 1999;104:807. Abstract 5.
2. Holzbeierlein J, Pope JC, Adams MC, et al. Urodynamic profile of myelodysplastic children with spinal closure in utero [abstract]. *Pediatrics*. 1999;104:811. Abstract 17.
3. Dodson J, Surer I, Baker L, Gearhart JP. The newborn exstrophy bladder too small for primary closure: evaluation, management and outcome [abstract]. *Pediatrics*. 1999;104:821. Abstract 42.
4. Grady RW, Mitchell ME. Complete primary repair of exstrophy. *J Urol*. 1999;162:1415-1420.
5. Coplen DE. Preliminary experience with neonatal complete primary closure of exstrophy/epispadias [abstract]. *Pediatrics*. 1999;104:822. Abstract 44.
6. Kropp BP, Cheng EY. Management of female bladder exstrophy/epispadias with total urogenital complex mobilization [abstract]. *Pediatrics*. 1999;104:822. Abstract 45.
7. Chan DY, Jeffs RD, Gearhart JP. Determinants of continence after bladder neck reconstruction in the bladder exstrophy population [abstract]. *Pediatrics*. 1999;104:823. Abstract 48.
8. Surer I, Baker LA, Jeffs RD, Gearhart JP. The modified Cantwell-Ransley repair in exstrophy and epispadias: ten years' experience [abstract]. *Pediatrics*. 1999;104:824. Abstract 49.
9. Park JM, Houck CS, Sethna NF, et al. Ketorolac suppresses postoperative bladder spasms after intravesical ureteral reimplantation [abstract]. *Pediatrics*. 1999;104:827. Abstract 55.
10. Casale AJ, Austin PF, Cain MP, et al. Nonpalpable undescended testis: does the order of procedure affect outcome and cost? A prospective randomized analysis of laparoscopy and groin exploration [abstract]. *Pediatrics*. 1999;104:829. Abstract 61.
11. Lee PA, Coughlin MT, Bellinger MF. Pretreatment testicular location: no difference in paternity of undescended testis after unilateral cryptorchidism [abstract]. *Pediatrics*. 1999;104:842. Abstract 94.
12. Hurwitz RS. How well does contralateral testis hypertrophy predict the absence of the nonpalpable testis? [abstract]. *Pediatrics*. 1999;104:844. Abstract 98.
13. Noe HN. Hypercalciuria and stone recurrence in pediatric urolithiasis [abstract]. *Pediatrics*. 1999;104:833. Abstract 73.
14. Ulman I, Jayanthi VR, Koff SA. The long term follow-up of newborn hydronephrosis initially managed nonoperatively [abstract]. *Pediatrics*. 1999;104:836. Abstract 78.
15. Kohler M, Wilcox D, Gordon I, et al. Primary obstructive megaureter managed by ureteric stenting: experience and long-term outcome [abstract]. *Pediatrics*. 1999;104:838. Abstract 84.